Cardiothoracic Transplantation (TX)

Clinical-Pathologic Conference

Historical Perspectives

Brief Communications

Cover Photograph

Chest deformities such as scoliosis and pectus excavatum are commonly associated with Marfan syndrome and can cause restrictive pulmonary dysfunction, although the connective tissue defects in Marfan syndrome seem to have little clinical effect on such dysfunction. Accordingly, patients with severe chest deformities are still predisposed to a risk of respiratory deterioration even after a successful extubation.

Three-dimensional computed tomographic scan revealed the dilated aortic root (62 mm in diameter) and severe scoliosis with marked elongation of the descending aorta along the vertebral bodies.[1] Preoperative spirometry showed a severe restrictive pattern: vital capacity, 1080 mL (28% of predicted value). Aortic root replacement was performed with a composite graft. Although respiratory distress developed immediately after the extubation, we managed to avoid reintubation with bilevel positive airway pressure ventilation. The patient was discharged from the hospital without any complication.

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Reference

1. Adachi I, Ogino H, Imanaka H, Matsuda H, Minatoya K, Sasaki H. Aortic root replacement in a patient with pulmonary dysfunction caused by severe chest deformity associated with Marfan syndrome. *J Thorac Cardiovasc Surg.* 2005;130:213-5.

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